Saudi Journal of Ophthalmology (2014) 28, 338–340

Case Report

Lacrimal gland CD5-positive, primary, extra-nodal marginal zone lymphoma of mucosa associated lymphoid tissue (MALT) – Type

Kaustubh Mulay, DNB [a,b,*]; Santosh G. Honavar, MS, FACS [c,d]

Abstract

Mucosa-associated lymphoid tissue (MALT) lymphoma of the ocular adnexae is rare. A 39-year-old woman presenting with proptosis was diagnosed to have non-Hodgkin's lymphoma with intermediate-sized cells and lymphoepithelial lesion. Unlike most MALT lymphomas, this lymphoma was found to be CD5-positive. Small lymphocytic lymphoma/chronic lymphocytic leukemia and lymphoplasmacytic lymphoma are two other entities that are CD5-positive and have a morphological pattern similar to MALT lymphoma. The case report and approach to the diagnosis is discussed.

Keywords: MALT, Lymphoma, CD5, Lacrimal gland

http://dx.doi.org/10.1016/j.sjopt.2014.04.004

Introduction

Mucosa-associated lymphoid tissue (MALT) – type marginal zone lymphoma (MZL) involves organs that normally lack lymphoid tissue. No specific immunohistochemical marker exists for MALT lymphomas. Usually known to be CD5-negative, they might rarely exhibit CD5-immunoreactivity.[1,2] We report on a 39-year-old woman with a CD5-positive MALT lymphoma of the lacrimal gland.

Case report

A 39-year-old woman presented with symptoms of swelling in the left eye and diplopia for 2 months (Fig. 1a). She was unaware of any systemic illness. On examination, she had 8-mm abaxial proptosis with 4 mm hypoglobus (Fig. 1b) and restriction of ocular movements in abduction and elevation. A firm mass was palpable in the superotemporal orbit in the region of the lacrimal gland. There were no signs of local inflammation. Schermer 1 values were 35 mm in 5 min in the right eye and 21 mm in the left eye. Rest of the ocular examination was unremarkable. Computed tomography showed an isodense diffuse mass involving the left lacrimal gland, extending laterally to involve the lateral rectus muscle with (Fig. 1c). Clinical diagnosis of lymphoproliferative lesion or nonspecific orbital inflammation was considered and a transperiosteal incisional biopsy was performed by a sub-brow anterior orbitotomy approach.

Microscopic examination showed a dense, neoplastic lymphoid infiltrate involving the lacrimal gland with splaying and partial destruction of the entrapped lacrimal gland tissue. Lymphoepithelial lesions with infiltration of acini by neoplastic cells were seen. The neoplastic lymphoid cells were intermediate-sized with scant cytoplasm and an irregular nucleus. Some cells had a plasmacytoid appearance.

Received 17 December 2013; accepted 16 April 2014; available online 21 June 2014.

[a] National Reporting Centre for Ophthalmic Pathology, Centre For Sight, Hyderabad, India
[b] Ocular Pathology Service, L.V. Prasad Eye Institute, Hyderabad, India
[c] Department of Oculoplastics and Ocular Oncology, L.V. Prasad Eye Institute, Hyderabad, India
[d] Department of Oculoplastics and Ocular Oncology, Centre For Sight, Hyderabad, India

* Corresponding author at: National Reporting Centre for Ophthalmic Pathology, Centre For Sight, Ashoka Capitol Building, Banjara Hills, Road No. 2, Hyderabad 500034, India.
e-mail address: kaustubh.m@hotmail.com (K. Mulay).

Peer review under responsibility of Saudi Ophthalmological Society, King Saud University

Production and hosting by Elsevier

Access this article online:
www.saudiophthaljournal.com
www.sciencedirect.com

Figure 1. (a and b) Abaxial proptosis and hypoglobus of left eye. (c) CT scan showing a homogenous mass in the left orbit in the region of lacrimal gland with extension to involve the lateral rectus.

Conspicuous nucleoli were seen in some cells (Fig. 2). The tumor cells expressed immunoreactivity for CD20, CD5 and Bcl-2 (Fig. 3) but were negative for CD10, CD23, CD30, TdT, Cyclin D1, Bcl-6 and CD3. A monotypical IgM expression was seen. Ki-67 labeling index was moderate up to a maximum of 15%. CD3-positive, reactive T-cells were seen in the background. Bone marrow and peripheral blood examination were normal. Systemic examination was normal. A diagnosis of CD5-positive MALT lymphoma was made. The patient was treated with 4000 cGy fractionated external beam radiotherapy to the left orbit. She is alive and well with complete regression at 5 months follow-up.

Discussion

Lymphomas involving the orbit and ocular adnexae are rare. MALT lymphoma appears to be the most frequent ocular adnexal lymphoma. MZLs are a group of indolent B-cell non-Hodgkin's lymphomas that arise from the marginal zone of lymphoid tissues. The World Health Organization (WHO) classifies them into 3 distinct types: splenic MZL, nodal MZL, and MALT lymphoma (or extranodal MZL). MALT lymphoma differs from its splenic and nodal counterparts as it arises in organs that normally lack lymphoid tissue (like stomach, lung, and salivary and lachrymal glands) but have accumulated B-cells in response to either chronic infections or autoimmune processes. Initially described to involve the stomach, MALT lymphoma has now been described to occur in the salivary gland, thyroid, intestine, lacrimal gland, conjunctiva and skin. MALT lymphomas are preceded by chronic antigenic stimulation and involve deregulation of the nuclear factor κB (NF-κB) pathway. Chlamydia psittaci is supposedly an important etiologic agent in the histogenesis of orbital MALT lymphoma.[3] MALT lymphomas are characterized by infiltration of neoplastic lymphoid cell between pre-existing follicles. The morphology of these neoplastic cells may vary within the same tumor. The tumor cells are of intermediate size with a scant cytoplasm and an irregular nucleus

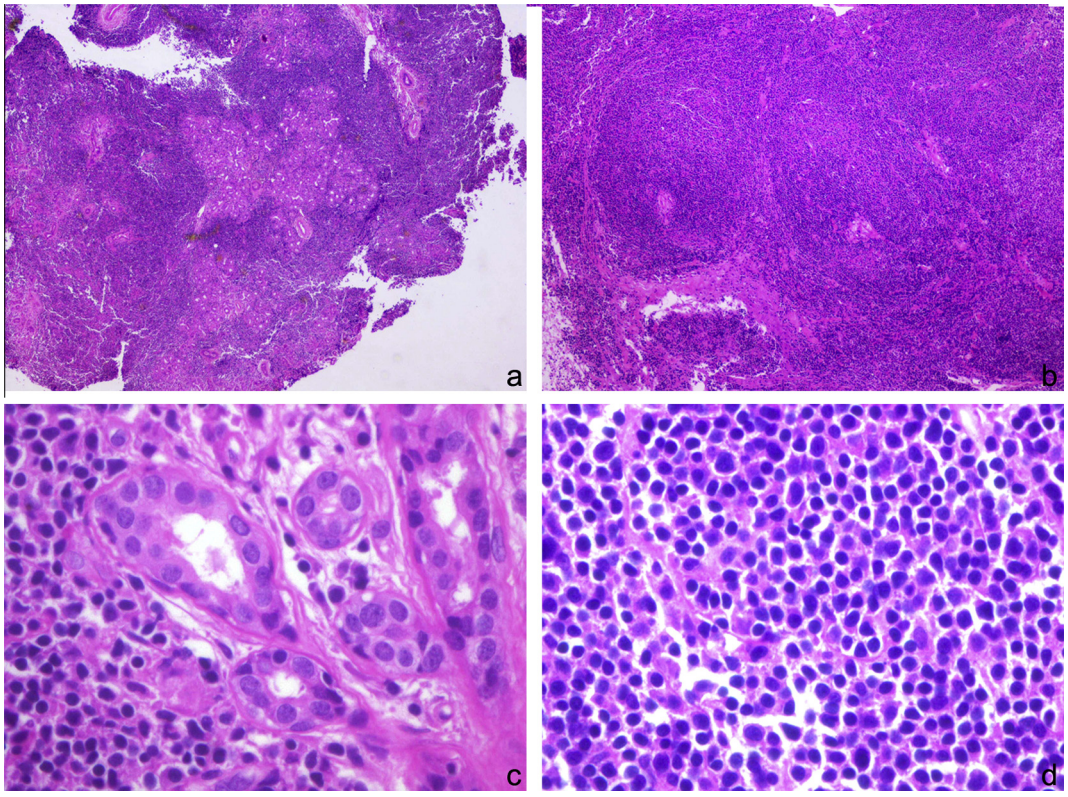

Figure 2. (a) Diffuse lymphoid tumor infiltrating and destroying the lacrimal gland (H and E ×20). (b) Lymphoid follicles with pale staining germinal centers (H and E ×40). (c) Infiltration of lacrimal acini with neoplastic lymphoid cells forming lymphoepithelial lesions (H and E ×400). (d) Intermediate-sized, centrocyte-like neoplastic lymphoid cells (H and E ×400).

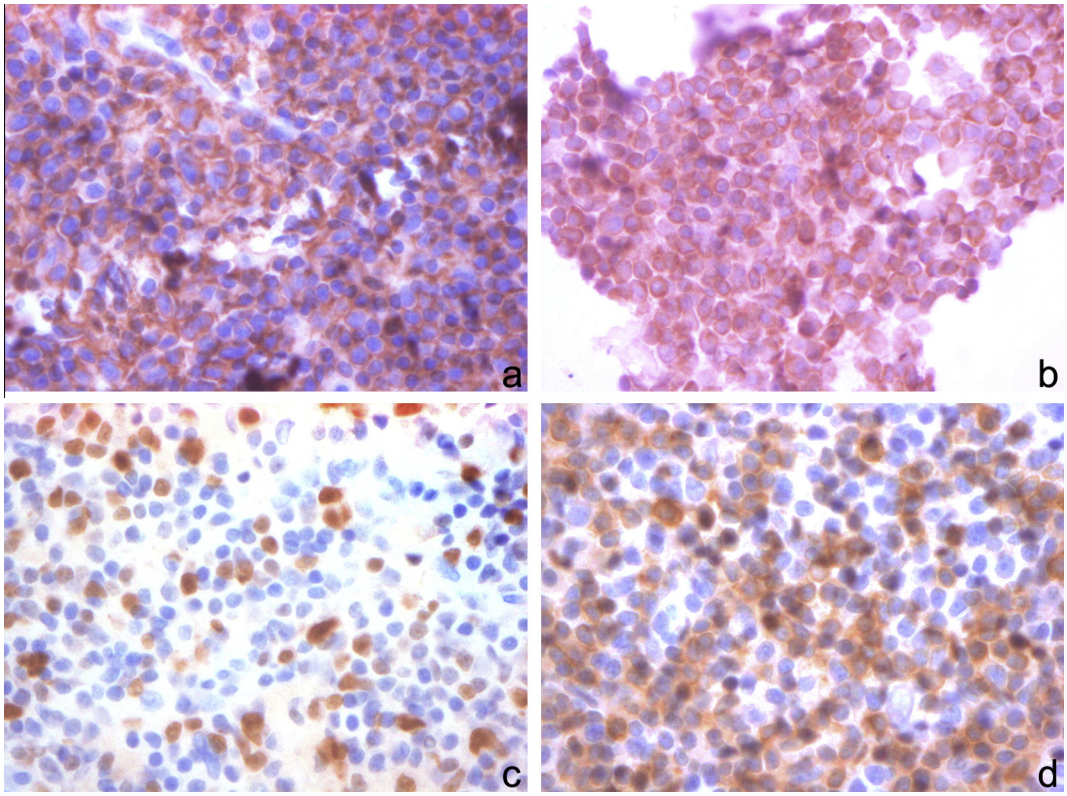

Figure 3. (a) Immunoreactivity of tumor cells for CD20 (×400). (b) Cytoplasmic immunoreactivity of tumor cells for Bcl-2 (×400). (c) Immunoreactivity for Ki-67 (×400) and (d) Immunoreactivity of tumor cells for CD5 (×400).

resembling centrocytes of the follicular center. Plasma cell differentiation is seen and may sometimes be prominent. The bona fide epithelial and mucosal injury, typified by the so-called ''lymphoepithelial lesion'', characterized by clustering of neoplastic cells infiltrating and destroying the glandular epithelium, helps diagnose MALT-lymphomas.[4] There is no specific marker for MALT-lymphoma. Immunohistochemically, the neoplastic MALT lymphoma cells co-express CD20 and surface light-chain restriction but are negative for CD10 and CD5. Rarely, CD5 immunoreactivity has been described in MALT lymphomas.[1,2] The role of CD5 expression in the prognosis of MALT-lymphomas is controversial.[5] Its aberrant expression was thought to be related to an aggressive behavior. However, reports of indolent CD5-positive MALT lymphomas exist in literature.[2] Lymphoblastic lymphoma (LPL) and small cell lymphoma/chronic lymphocytic leukemia (SLL/CLL) are other CD5-positive, B-cell lymphoproliferative disorders that share morphology similar to MALT lymphomas. Absence of immunoexpression for CD23, as seen in our case, would rule out a SLL which is almost invariably CD23-positive. LPL may have a CD23-negative immunophenotype similar to MALT lymphoma. A thorough systemic examination would help in this case. LPL and SLL/CLL are systemic diseases while MALT lymphoma is localized to extranodal regions.

MALT lymphoma is a B-cell lymphoproliferative disease that is usually CD5-negative but may rarely express a CD5-immunoreactivity. It being a localized disease of extranodal sites, rarely shows systemic involvement, and can be treated with local excision or external beam radiotherapy. Hence, its distinction from other CD5-positive lymphomas with a similar morphology becomes important.

Conflict of interest

The authors report no conflicts of interest in this work.

References

1. Jaso J, Chen L, Li S, et al. CD5-positive mucosa-associated lymphoid tissue (MALT) lymphoma: a clinicopathologic study of 14 cases. *Hum Pathol* 2012;**43**:1436–43.
2. Wenzel C, Dieckmann K, Fiebiger W, Mannhalter C, Chott A, Raderer M. CD5 expression in a lymphoma of the mucosa-associated lymphoid tissue (MALT)-type as a marker for early dissemination and aggressive clinical behaviour. *Leuk Lymphoma* 2001;**42**:823–9.
3. Ferreri AJM, Dolcetti R, Magnino S, Doglioni C, Ponzoni P. Chlamydial infection: the link with ocular adnexal lymphomas. *Nat Rev Clin Oncol* 2009;**6**:658–69.
4. Katić V, Katić K, Vučić M, Gligorijević J, Dimov D, Zlatić A. The histopathology and immunohistology of gastric MALT lymphoma. *Arch Oncol* 2004;**12**(Suppl. 1):5–6.
5. Harris NL, Jaffe ES, Stein H, et al. A revised European–American classification of lymphoid neoplasms: a proposal from the International Lymphoma Study Group. *Blood* 1994;**84**:1361–92.